

Palilalia, Paligraphia and Progressive Non fluent Aphasia in a patient with Progressive Supranuclear Palsy

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ABSTRACT

Recent studies have demonstrated that a subgroup of patients with progressive supranuclear palsy (PSP) may be associated with progressive apraxia of speech (AOS), nonfluent aphasia (PNFA), or both. We describe the cognitive-linguistic characteristics of patient with progressive supranuclear palsy with co-occurring progressive non fluent aphasia, palilalia, paligraphia and obsessive compulsive disorder symptoms. He presented with history of behavioral disturbance for six years and progressive difficulty in speaking for the last two years. Behavioral abnormalities were manifested in the form of aggressiveness, suspicion towards wife, using abusive language, apathy, withdrawn behavior, impairment in short term memory, sleep disturbances, tactile hallucinations and repetition of the same action. Spontaneous speech was characterized by significant palilalia and writing was characterized by paligraphia which had worsened over the last two years. Auditory comprehension and naming was good. He also demonstrated extrapyramidal features and mild bradykinesia.

Key words: Progressive supranuclear palsy, Progressive nonfluent aphasia, Palilalia, Paligraphia

INTRODUCTION

There have been few descriptions of the association of progressive supranuclear palsy (PSP) with progressive nonfluent aphasia (PNFA) and apraxia of speech [1-5]. Karnik et al [6] described a 64-year-old woman with frontotemporal lobar degeneration, progressive non-fluent aphasia, obsessive compulsive disorder (OCD) and progressive supranuclear palsy. There is no reported evidence of the occurrence of palilalia and paligraphia with PSP. Critchley [7] was the first to report palilalia in five patients with post encephalitic Parkinsonism, one patient with pseudobulbar palsy and in another patient with emotional lability. Palilalia and paligraphia are also reported in patients post road traffic accident and following cerebrovascular accident [8-10]. It was also reported in a patient with idiopathic Parkinsons disease [11] and in 15 out of 24

patients with advanced Parkinson's disease [10]. Palilalia is generally associated with bilateral basal ganglia lesion and Parkinson's disease [12], Tourette syndrome [13,14], progressive supranuclear palsy [15,16], right hemisphere damage [17]. Literature also reports of neuroleptic-induced palilalia (Garcia, 1990). There is sparse literature on the progression of speech and language features in PSP patients having PNFA [19]. The co-occurrence of progressive supranuclear palsy, nonfluent aphasia, OCD, palilalia and paligraphia is rare and we report such a case highlighting the speech-language findings.

CASE HISTORY

Mr. MR. is a 51-year old male with history of behavioral disturbances and progressive difficulty in speaking since 49 years of age. Behavioral issues started as aggressiveness, and suspicion and of his

wife. He was apathic and withdrawn for the last two years. He also had progressive difficulty in speech characterized by compulsive repetition of syllables and words and subtle difficulty in word retrieval. Writing was characterized by paligraha and micrographia. Since 5 months prior to admission his sibling noticed impaired attention, repeating same action or words, difficulty in remembering recent events, inappropriate smiling, muttering, compulsive face washing and bathing. He started eating at normal rate but later on ate very fast and often spilled food. He also presented with altered sleep wake cycle and singing, laughing and muttering to self. He had slowness of movements, reduced facial expressions and mild tremors in hands since last six months. Difficulties

in writing, reading, language and behavioral issues lead to loss of job, reduced social interaction and inability to participate in leisure activities. The onset and course of illness was ascertained from his brother with whom he was currently staying.

Evaluation

Neurological examination six years after onset of behavioral issues and 2 years after onset of speech problem revealed relatively young onset speech disturbance, perseveration, palilalia, naming impairment, relatively preserved comprehension, delusion, aggression, emotional lability, apathy, extrapyramidal features and mild bradykinesia. Fig 1. represents writing sample of the patient demonstrating Paligraha and micrographia

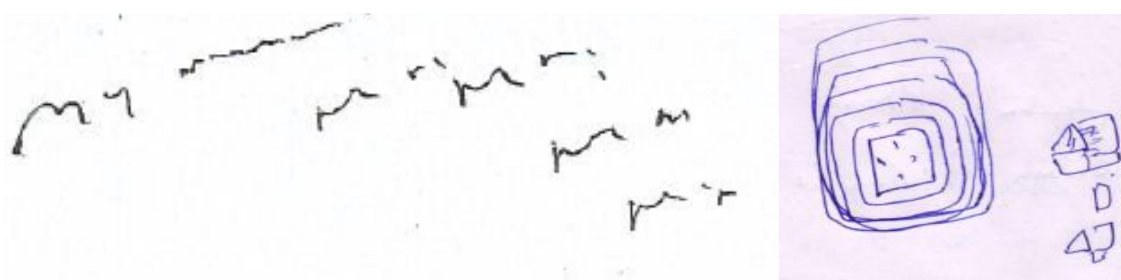


Fig 1: writing sample of the patient with paligraha and micrographia

Basic ADLs were good, but instrumental activities of daily living comprising of telephone usage, managing money, going out independently and leisure time activities were affected. The UPDRS III Speech score in the medication OFF stage was 3, characterized by marked impairment and difficulty to understand speech.

There was no apraxia (verbal or nonverbal). He had impaired fronto temporal functions, severe impairment of response inhibition with significant speech disturbance. MRI showed significant frontal and anterior temporal atrophy with more asymmetry on left side. It also indicated perisylvian, cingulate asymmetry with L>R atrophy consistent with PNFA. Minimal state score was 23/30. The patient was diagnosed as progressive extrapyramidal

disorder with progressive nonfluent aphasia (PNFA) variant of frontotemporal dementia. It was opined that it may persist as PNFA or may evolve into progressive supranuclear palsy with or without apraxia of speech.

Speech and language evaluation

Speech and language manifestations consisted of a progressive course in the last 3 months as described by the patient and his brother. On examination the patient presented with compulsive repetitions of syllables (30 to 40 times and sometimes more than 100 reiterations) in spontaneous speech, naming and responsive speech (propositional tasks) with occasional repetition of words and phrases. Reiterations were also seen while counting, reciting names of months and to a lesser extent while reciting prayer (non propositional

tasks). It was accompanied by reduced loudness and increased rate and were reduced when the patient was less anxious and when attention was focused on speech. The disfluencies were more characteristic of

palilalia (Figure 2). The speech samples were audio recorded using a Sony MZ-55 digital recorder and were transferred to Computerized Speech Laboratory (CSL-4500) at a sampling rate of 44.1 kHz.

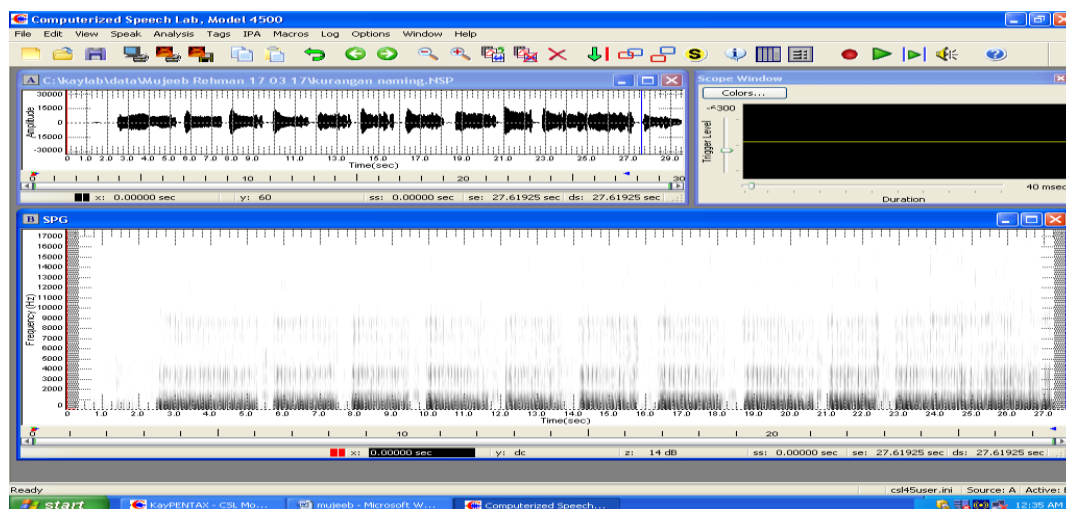


Figure 2: Spectrogram of the word /kurangan/.

The word was completed in a total of 23 seconds. There were 9 syllable trains each with approximately 12 reiterations of phoneme /k/ before producing the complete word (Total 108 reiterations in 20 seconds). It is noticeable that the first syllable train of /k/ was for 2 sec, 2nd to 8th syllable train of /k/ was for 1.5 seconds, 9th syllable train was for 1 sec. The complete word was uttered in 4 seconds (between the 23rd and 27th second).

The percentage of reiterations in a 256 word passage in reading was 35.56% (91 on 256) and in sentence repetition was 20.31% (52 on 256). The reiterations were the same in the medication ON and OFF states. No family history of stuttering was reported. As a result of his increasingly unintelligible speech Mr.M.R avoided conversations and other social interactions and the speech difficulty had rendered him unable to continue his job.

No significant dysarthria or swallowing difficulty was reported. There were no inconsistent responses, phoneme distortions or metathesis suggestive of apraxia of speech, except for minimal groping while saying multisyllabic utterances. Spontaneous speech was

significantly more disfluent than oral reading, repetition and singing. He could read fairly well without much dysfluencies. He had occasional echolalia. Writing was characterized by similar errors as in speaking with involuntary repetitions and difficulty in terminating writing (paligraha and micrographia), with repetitive writing of the first syllable of the word, word and phrase reiterations. He had fair semantic association scores in the Indian semantic battery (26 on 42), good naming in the picture naming test (38/40). He had good category sorting abilities, but performed poor on category fluency. His poor category fluency performance however has to be interpreted in view of his severe palilalia.

In the Western Aphasia Battery (WAB; Kertesz, 1980) he had a score of 7 in information content (correct responses to 4 of 6 items), 2 in fluency (single words and effortful, agrammatic and telegraphic). He scored 54 on 60 for yes/no questions and auditory word recognition, 30 on 80 for sequential commands, 42 on 100 for repetition, and 59 on 100 for naming. His aphasia quotient was 6.9 and cortical quotient was 13.8. These scores were indicative of transcortical motor aphasia. In

March 2017, his speech and language deficits worsened with clear signs of nonfluent progressive aphasia, mild agrammatism in reading, difficulties in writing and repetition. He made errors (distortions, metathesis etc) while repeating longer sentences. He also exhibited deficits in understanding complex grammatical sentences.

The oral peripheral mechanism evaluation based on Frenchay Dysarthria Assessment [20], revealed no difficulty in reflex, respiration, lip, jaw and palatal function. He had only limited change in volume. His voice deteriorated with the length of speech. The movement of lips in conversation was minimal with poor acoustic representation. The findings were suggestive of mild hypokinetic dysarthria. Based on the above observations and findings he was diagnosed to have palilalia, paligraha and progressive nonfluent aphasia secondary to progressive supranuclear palsy. He was advised to undergo neuropsychological evaluation, cognitive-linguistic stimulation, monitoring and pacing of speech. He was also counselled regarding the use of alternative and augmentative communication. The patient was advised speech therapy and he attended the same twice weekly for 2 weeks. The goals were to reduce his speech rate and reduce the number of palilalic utterances using pacing board and hand tapping techniques. Though these external cues were useful for few seconds, speech coordination could not be maintained for spontaneous speech, it was characterized by significant palilalia. Hence therapy was refocused on improving his functional speech by using alternative and augmentative modes for word and sentence level communication (text-to-speech) and communication strategies for care giver. He was advised by the neurologist to undergo repetitive transcranial stimulation for reducing the palilalia.

DISCUSSION

We report the clinical findings of a case who presented with features of nonfluent aphasia, palilalia, paligraha and obsessive-compulsive disorder (OCD). Expressive language was agrammatic, auditory comprehension and naming was good and he had relatively preserved semantic and cognitive functions. The patient was classified as progressive nonfluent aphasia according to the FTLN international criteria [21]. The patient also had minimal hypokinetic dysarthria. The pathological features were suggestive of progressive supranuclear palsy (PSP). There is very little information in the literature about the evolution of extrapyramidal features in PSP, especially when it presents initially as PNFA.

Our patient presented with behavioral issues and progressive difficulty in language expression with characteristic features of progressive nonfluent aphasia (PNFA). The association of PSP and PNFA has been reported by Boeve et al. [1] and Mochizuki et al. [2]. Boeve et al. [1] reported of 71-year-old man with anomia and grammatical errors. He initially had symptoms of Parkinsonism and AOS. His language deficits and AOS worsened after 3 years and pathological examination confirmed PSP. They noticed that he did not have any typical features of PSP. Mochizuki et al. [2] reported of a 64-year-old man with reduced expressive language and preserved auditory comprehension in the absence of PSP or Parkinsonian symptoms. He had paraphasias at 67 years, and dysphagia and postural instability at 73 years. Pathological examination later revealed PSP. Karnik et al. [6] reported of a 64-year-old woman with OCD and effortful speech who was later confirmed to have PSP. Josephs et al. (2006) reported that 10% of patients with PSP had associated AOS and PNFA in their cohort of 100 autopsy-confirmed cases. They opined that the extrapyramidal features may be seen in the later stages of PSP and may not be as severe as in typical PSP. Dysarthria (hypokinetic) was minimal

in our patient along with mild extrapyramidal symptoms.

Palilalia can be described as involuntary repetitions of phonemes, syllables, words or phrases just uttered by the patient. Our patient had reiterations ranged from 56 to 108 phoneme and syllable repetitions in spontaneous speech. Palilalia may also be characterized by reduced volume and intelligibility towards the end of repetitions in a repetition train and may even become aphonic (palilalie aphone). Some studies have reported an increasing rate of repetition of words and phrases [22], akin to the festinating gait in patients with parkinson's disease [23]. John Van Borsel et al. [11] opined that the durational variability in palilalia may be indicative of a variable motor program. They reported more fade-in repetitions (repetition of part of word or phrase before uttering the word or phrase; suggestive of difficulty in initiation of speech) in automatic speech tasks and fade-out reiterations (suggestive of speech inhibition problems) in propositional speech in their patients with motor vehicle accident and cerebrovascular disease. Kent et al. [24] also reported fade-in repetitions in their patient with palilalia. The findings for our patient are suggestive of problems with initiation and progression of speech as is evident from the fade-in repetitions, which was more pronounced in propositional tasks. No actual durational measurements were made in our study, but perceptually an increasing rate of reiterations could be perceived. Kent et al. [24] opined that among the subgroups of patients with palilalia, an increasing rate may be indicative of extrapyramidal involvement, a constant rate suggestive of an invariant motor program and temporal variability and 'expanded structure repetition' or repetition of progressively larger units indicative of involvement of new motor instructions. MRI of our patient was suggestive of extrapyramidal and frontotemporal involvement.

A motor and cognitive hypothesis has been proposed to understand the

underlying mechanisms of palilalia. The motor hypothesis has equated the reiterations to freezing episodes seen in Parkinsons disease. They said that both the disorders have similar characteristics with difficulty in inhibiting or shifting from one movement to next [25]. Freezing is also accompanied by an increase in frequency and reduced amplitude as movement progresses [26]. Cognitive hypothesis for reiterations by Levelt [27] and Ackermann and Ziegler [28] claimed a deficit at prearticulatory level. The fact that the iterations are more in propositional than nonpropositional speech and the frequent association of word finding difficulties with reiterations (with reiterations acting as fillers till the complex word is uttered) and increase in reiterations with increased linguistic demands are supportive of the deficit at the prearticulatory level. Speech characteristics in our patient showed involvement at the motor as well as cognitive level.

Alfredo Ardila et al. [9] noticed speech task based differences in the severity of palilalia, with palilalia being present in spontaneous speech alone compared to verbal fluency and naming tasks. They discussed that this may be due to the different brain regions underlying the production of these tasks; with spontaneous speech being modulated mainly by left frontal areas [29] and verbal fluency and naming by left temporal areas [30]. There are mixed opinions regarding the effect of propositional and nonpropositional speech on severity of palilalia. Kent et al. [24] proposed a clear distinction by stating that propositional speech (conversation, naming) is affected more severely than nonpropositional speech (reading aloud, reciting verse or prayer, telling days of week, months of year, letters of alphabet, counting). Benke and Butterworth [31] stated that palilalia was similar in both tasks. Spontaneous speech was spared in patients with palilalia [12], whereas repetition was affected. In our patient, the reiterations were

seen more in propositional than automatic speech tasks.

There have been varying contentions with respect to the locus of reiteration and effect of phonological factors on palilalia. Lebrun [32] reported that patients with palilalia repeated words and phrases as opposed to phoneme and syllable repetition in developmental stutterers. Van Borsel, Van Coster, & Van Lierde [33] found that words and phrases in the final position were more often repeated in contradistinction to the dysfluencies seen in patients with developmental stuttering. In contrast, Lapointe and Horner [34] reported that 55% of reiterations were seen in the beginning of a sentence. They also reported differences in palilalia severity based on phonological environment with words starting with consonant clusters, plosives, affricates and labiodentals most affected and those with vowels and linguadentals least affected. The reiterations were mainly phoneme and syllable repetitions in our patient and were seen at any loci of the sentence. Reiterations were seen irrespective of the type of vowels or consonants.

Palilalia co-occurred with paligraphy in our patient. The patterns of reiterations were similar in spoken as well as written language. Ardila et al. [9] reported a case of severe palilalia with transient paligraphy and speculated that the co-occurrence of both symptoms may be indicative of a broader disorder than peripheral speech defect alone. Palilalia was more pronounced than paligraphian in their patient and there was no paligraphy after three weeks.

Our patient did not have AOS or phonological errors, but did show distortions and metathetic errors with increased length of the sentence in the repetition task. Increasing errors with sentence length may be ascribed to the demands of auditory processing on the sensorimotor aspects of speech motor control in patients with AOS [35]. Mr.MR had minimal hypokinetic dysarthria. Hypokinetic dysarthria was reported only in

11 out of 18 people with nonfluent PPA and AOS [36].

Management

Management strategies included caregiver education regarding the evolving disease process and functional deficits and the associated neurological, neuropsychiatric and speech-language and cognitive decline. They were also explained about communication strategies, cognitive-linguistic stimulation and pacing for reducing speech rate. They were also advised regarding low technology alternative and augmentative communication (AAC) (communication books, communication boards and cards, pantomime and gesture), and other high-tech AAC devices (text-to-speech software). They were also advised about informing relatives and close friends regarding providing enough time to the patient for communication.

CONCLUSION

The present study was undertaken to describe the characteristics of palilalia and paligraphy in a patient with PNFA and PSP. The findings of the present study suggest that it may be more appropriate to define palilalia as reiterations that are indicative of problems with the initiation of words and phrases. It also indicates that the severity of palilalia may vary with respect to proportionality and length of the utterances. The results have to be viewed by keeping in mind that palilalia can be subjected to increased interindividual variability. It is also not yet clear if palilalia shows a constant pattern over time. Changes in the pattern of palilalia under the influence of medication also needs to be investigated.

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